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PROBLEMS OF THE PATHOGENESIS OF THE UNOBSTRUCTED FORM OF HYDROCEPHALUS IN CHILDREN

[Following is the translation of an article by B. N. Klosovskiy, B. V. Lebedev, V. R. Purin entitled Voprosy Patogeneza Otkrytoy Formy Gidrotsefalii u Detey (English version above), in Voprosy Neyrokhirurgii (Problems of Neurosurgery), Vol. XXIV, No. 3, Moscow, 1960, pages 54-57.]

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In this work we should like to direct attention to a distinctive complex of symptoms in children which is expressed in an increase of intracranial pressure, dilatation of the ventricles and subarachnoid spaces against the background of complete patency of the spinal fluid communications. This symptom complex is encountered in children quite often and therefore deserves special description.

In a period of six years there were 160 children in the neuropsychiatric clinic of the Institute of Pediatrics of the Academy of Medical Sciences USSR with organic lesions of the central nervous system,

in which, in the absence of general infections, intoxications or meningoencephalitis, the clinical characteristics of these children amounted primarily to mental retardation, at times very much pronounced. They had diffuse and focal lesions of the nervous system in the form of pareses and paralyses, athetosis, spasms, etc. In all cases, the children had information in their history concerning a pathological course of pregnancy in the mother as a result of endocrine diseases, vitamin deficiency, anemia, the exhaustion of her chemical and mechanical forces for expelling the fetus or a history in which the child had suffered birth trauma or asphyxia at birth.

In 54 children the spinal fluid pressure in a position of lying on the side under conditions of a calm wakeful state was within limits of 200-300 millimeters of water; in 49 children, within limits of 300-400 millimeters; in 51 children, within limits of 400-500 millimeters, and in six children the intracranial pressure was higher than 500 millimeters of water. The fluid was colorless, the cell count did not exceed eight cells per cubic millimeter. The protein content was always decreased (0.033-0.066 per thousand).

The pneumoencephalographic investigation was performed in all children. It showed complete patency of the ventricular system and of the subarachnoid spaces. In the majority of children a dilatation of the ventricles was seen on the pneumoencephalogram. In their lateral

ventricles the anterior horns and the middle portion were always dilated to a much greater degree than the posterior and inferior horns. Not uncommonly, there were inlet-like dilatations of the lateral ventricles. In the frontal projection the lumen of one ventricle was larger, frequently, and had a different shape from that of the opposite lateral ventricle. The third ventricle, which normally has the shape of a narrow slit, was oval or even rounded, evidently because of a bilateral atrophy or hypoplasia of the thalamus. Dilatation of the aqueduct of Sylvius was distinct, but rarely reached a considerable degree. The fourth ventricle showed a very moderate dilatation. The subarachnoid space was well filled with air. However, here, instead of a delicate reticular pattern we noted an accumulation of air along the course of sulci. In the area of the frontal and parietal lobes the accumulation of air was of a cystic nature; not uncommonly, the subarachnoid space was generally dilated here. The cisterns at the base of the brain were always dilated.

Let us present some case histories.

1. Serezha M., age one year and six months. He was admitted to the clinic for a lag in physical and mental development. His parents were healthy; pregnancy in the mother had been normal. Twins were delivered in the seventh month; the mother had weak parturient activity. The boy was born first, weighed 2400 grams, cried immediately but weakly.

The second child was a girl weighing 1800 grams; she died after several days, as a result of cerebral hemorrhage at the time of delivery. In the lying-in home the boy was in a serious condition, and began to suck on the fourth day. The following diagnosis was made: birth trauma, hemorrhage into the brain.

At the time of admission the following were found: weight, 8040 grams; height, 75 centimeters; circumference of the head, 47 centimeters. The anterior fontanelle was 2x2 centimeters. He did not hold his head up, did not sit nor support himself on his feet. He could fixate his gaze, smiled, reacted poorly to toys, and said nothing. There was a convergent strabismus on the right, flattening of the right nasolabial fold. There was an increase in tone in the arm flexors, more on the right, and of the abductor muscles of the thighs. The knee reflexes were considerably increased. There was a pronounced athetosis of the distal portions of the arms and of the musculature of facial expression. There was a general hyperesthesia.

The intracranial pressure was 300 millimeters of mercury. The cell count in the spinal fluid was three; the protein was 0.066 per thousand; the Wasserman reaction was negative. On the pneumoencephalogram -- the lateral ventricles were dilated and somewhat deformed. There was cystic dilatations of the subarachnoid spaces, chiefly in the frontal areas.

2. Vasya M., age eight months. Was admitted because of a lag in psychophysiological development. The parents were healthy. The mother's pregnancy had proceeded satisfactorily. Delivery was at term and lasted about three days. A day after discharge of the amniotic fluid the child was extracted by Caesarian section. His weight was 3700 grams. He did not cry immediately.

At the time of admission the following were found: weight, 7330 grams; height, 70 centimeters; circumference of the head, 46.5 centimeters. He swallowed poorly. He reacted weakly to his surroundings, did not fixate his gaze, could not hold his head up, did not see independently and could not support himself on his feet. The muscle tone was increased in the hands and feet. There was athetosis of the facial musculature and of the hands. The optic fundus was unchanged. The intracranial pressure was 250 millimeters of water. The cell count in the spinal fluid was eight cells per cubic millimeters; the protein was 0.1 per thousand. The Pandy and Wasserman reactions were negative. On pneumoencephalography there was a dilatation and deformation of the lateral cerebral ventricles, cyst-like dilatations of the subarachnoid spaces, particularly in the frontal, parietal and occipital areas.

An increase in the intracranial pressure, dilatation of the ventricles and subarachnoid spaces of the brain is what we characterize as a symptom-complex of unobstructed hydrocephalus. Here,

hydrocephalus is of an active nature and thereby is markedly different from those residual signs where dilatation of the ventricles and subarachnoid spaces is noted against the background of a normal or reduced intracranial pressure and, therefore, hydrocephalus has already been completed as a process (hydrocephalus ex vacuo). Therefore, in the cases which we have described we consider the main phenomenon to be increased intracranial pressure.

What is the origin of the unobstructed hydrocephalus in the children whom we examined with the consequences of disturbances in intrauterine development and with consequences of birth trauma and asphyxia? It is more convenient to begin a discussion of this with the analysis of the possible mechanisms of disturbance of circulation of the spinal fluid in children who have sustained birth trauma.

As is well known, birth trauma of the brain is expressed in injury of the sinuses, of the blood vessels of the brain surface, in the entry of blood into the subarachnoid space, in traumatization of the brain matter with rupture of blood vessels and the formation of hemorrhages in various parts of the brain, subcortical nuclei and brain stem. It is customarily considered that when blood enters the subarachnoid space an aseptic meningitis develops with a subsequent fibrosis of the pia mater, and sometimes with obliteration of the subarachnoid spaces.

These factors are considered the cause of disturbance in the spinal fluid circulation and of a change in the neurological status of the child subsequently (Bagley, 1928; Parker and Lehman, 1936; Sharpe and Maclaire, 1924). Important significance in the origin of hydrocephalus is ascribed also to a marked increase in the osmotic pressure of the spinal fluid when blood enters it (Howe, 1929).

In the literature, we have been unable to find any works, however, in which the changes in intracranial pressure have been studied in the late periods after subarachnoid hemorrhages. The data, which deal with adults, show that here, despite a certain reaction of the meninges (cystic arachnoiditis), the intracranial pressure is even reduced after the acute signs subside (S. V. Gol'man, 1949).

In our investigations we have imitated birth trauma, destroying the longitudinal sinus over a considerable extent in three-four-day old puppies and part of the convex surface of the cerebrum. In addition, the animals were subjected to sublethal asphyxia. Here, blood entered the subarachnoid space in a large quantity. However, by measuring the intracranial pressure in the animals one to five months after the procedure, we could not in any case find any increase in it (V. R. Purin). This makes us take a sceptical attitude toward the idea that increased pressure in the spinal fluid in the children whom we examined with the consequences of birth trauma and asphyxia occurred as a result

of the entrance of blood into the subarachnoid space.

In patients whom we observed, it was impossible to suspect any hypersecretion of the spinal fluid. Cases of hydrocephalus described in the literature, which occurred as the result of increased spinal fluid formation, are solitary. Here, the main cause of hypersecretion of the spinal fluid was shown at autopsy to be a tumor-like proliferation of the vascular plexuses of the brain (Davis, 1924). Recently, communications have appeared which make the hypersecretory nature of hydrocephalus in papillomas of the vascular plexuses doubtful, because, on the one hand, there is no proof of increased spinal fluid production by the papilloma; on the other hand, the secretion which it produces exerts an irritative effect on the ependyma and can lead to a granular ependymitis with subsequent obstruction of the aqueduct of Sylvius or to an adhesive process in the cisterns of the base of the brain (Russel, 1954). It is important to note the fact that in children with the late consequences of brain injuries at early stages of development atrophic changes in the vascular plexuses are found (B. N. Klosovskiy and M. Ya. Turetskiy). Therefore, it is more correct to look for the cause of increase in intracranial pressure in the children in whom we investigated in a disturbance in the spinal fluid resorption.

Current investigations on the development of the spinal fluid system in the course of ontogeny and of its activity in adults show

that the spinal fluid penetrates into the brain matter from the cerebral ventricle and submeningeal spaces, comes into contact with the brain elements, and is resorbed afterwards by a powerful venous segment of the vascular-capillary network of the brain.

During the intrauterine period, and in the child a certain time after birth the spinal fluid provides the brain with special protein substances which contribute to its rapid growth. Afterwards, losing the trophic functions, the spinal fluid dissolves the metabolic products of nerve tissue with its own enzymes, contributing to their excretion into the blood. It has been shown also that the passage of spinal fluid through the brain substance is not a simple impregnation but rather represents an active process brought about by the activity of the brain matter (B. N. Klosovskiy, 1947; V. R. Purin, 1958; N. S. Volzhina, 1960).

According to the observations of B. N. Klosovskiy (1947), the condition of the vegetative nervous system exerts a great influence on the level of intracranial pressure. Thus, for example in the cases where the brain swells up it is possible to cause a reduction in the intracranial pressure by the intravenous injection of glucose with ephedrin. The latter possesses an excitatory effect on the sympathetic nervous system. In experiments on animals a rapid reduction in the intracranial pressure was achieved by stimulation of the sympathetic nerve in the neck (B. N. Klosovskiy, 1944). Here, a certain narrowing of the arteries and

dilatation of the veins on the brain surface is observed, because of which better conditions are created for the absorption of spinal fluid. V. R. Purin (1958) in experiments on animals established the fact that during the active state of the brain, particularly during excitation, the spinal fluid is actively absorbed by the brain substance and is rapidly excreted from it into the blood. Correspondingly, the intracranial pressure in these cases is low. On the other hand, during sleep the activity of the absorption processes of the spinal fluid decreases sharply, and the intracranial pressure is increased. The activity of the spinal fluid system is directed, therefore, toward a satisfaction of the requirements of the brain metabolism, and the conditions of absorption of the spinal fluid are determined by the nature of activity of the nervous tissue.

On examination and everyday observation of the children with the late consequences of brain injuries at the early stages of development we were always struck by the inactivity of the majority of the children and their areactivity. They were not actively awake, a condition which is so characteristic of the normally developing child. In those cases where the behavior of the children was restless, their excitement was more of a subcortical nature. This undoubtedly is the result of the low level of metabolism of the nerve cells of the cerebral cortex.

The cause of these metabolic disturbances and reflex

brain activity in cases which we have are describing were disturbances in the cerebral development as a result of the effect on it of injurious factors during pregnancy, and in children who have sustained birth trauma, evidently, injuries to the nuclei of the brain stem, which regulate the circulation in the brain and its water metabolism. The latter supposition is confirmed by B. N. Klosovskiy's data concerning the change in the cerebral vascular tone as well as swelling of the brain with stimulation of certain brain stem nuclei and of the hypothalamus. These observations are confirmed now by many investigators (I. A. Prikhodchenko, 1955; Yu. N. Kvitnitskiy-Ryzhov, 1955 and others).

Based both on the clinical and experimental data we believe that specifically the low level of activity of the nerve cells of the brain to a considerable degree is responsible for the persistent increase in intracranial pressure in the children under our observation. Such relationships have been noted also in other conditions. For example, in congenital myxedema in children a high intracranial pressure has always been noted. This contributes to a low level of brain metabolism. After treatment with thyroid and aloe extract, simultaneously with the activation of reflex cerebral activity and activation of the metabolic processes the intracranial pressure is reduced (M. F. Yankova, 1960).

We must suppose that at first the increase in intracranial pressure is adequate for the particular functional condition of the

nerve cells; the conditions of increased intracranial pressure are just as adequate and necessary for the occurrence of metabolic processes in the nerve cells during sleep. However, afterwards, specficially as a result of its chronic nature the increase in intracranial pressure can lead to a retardation in cerebral development and a partial atrophy of the brain substance. This may be observed also experimentally (Z. N. Kiseleva, 1951; N. S. Volzhina, 1952). In our opinion, therefore, dilatation of the subarachnoid space, dilatation of the ventricles, that is, hydrocephalus, has a secondary origin, being the result of a chronic increase in intracranial pressure.

It is well known that the parts of the brain in which the formative processes occur actively are more vulnerable. Here, the predominant involvement of frontal and parietal lobes in the case of unobstructed hydrocephalus in children can be explained. The parietal, and particularly the frontal, lobes belong to the parts of the brain which mature late. In them the processes of myelinization, dendrite formation and the creation of the blood vessel architectonics occur for a longer time. Naturally, in the postnatal period they are more sensitive to the harmful influence of high intracranial pressure.

It is very important to note further that, as the experimental research of Z. N. Kiseleva and N. S.Volzhina has shown (1951, 1952), elimination of increased intracranial pressure, if we are dealing with young animals, leads to the activation of compensatory processes in

the brain and, particularly, to a considerable activation of the growth of capillaries in the cerebral cortex.

Based on the idea presented of the pathogenesis of unobstructed hydrocephalus in children with the late consequences of cerebral injury at early stages of development we are striving for the attainment of the following goals in the course of treatment:

1) To increase the level of metabolism and excitability of the nerve cells of the brain by means of drugs, vitamin therapy and a combination of routine and training measures; the latter of necessity includes stimulation of the activity of analyzers and chiefly of the vestibular analyzer;

2) to reduce the intracranial pressure by means of taps and dehydration therapy; the combined injection of hypertonic solutions and ephedrin which possesses a sympathicotropic effect is expedient; recently, we have been using phonurite (diamox) for this purpose;

3) to use aloe extract injections as additional therapy for the children; this stimulates the activity of nerve cells and the growth of brain capillaries.

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1288 END

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